FACTSHEET

Veterinary Services

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Animal and Plant Health Inspection Service

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Scrapie

Scrapie is a fatal, degenerative disease affecting the central nervous system of sheep and goats. Scrapie has had a significant impact on the sheep industry and has caused financial losses to sheep producers across the country.

In the United States, scrapie primarily has been reported in the Suffolk breed. It also has been diagnosed in a Border Leicester, Cheviots, Corriedales, a Cotswold, Dorsets, Finn sheep, Hampshires, Merinos, Montadales, Rambouillets, Shropshires, Southdowns, and a number of crossbreeds. Through August 1999, seven cases of scrapie have been reported in goats.

History

First recognized as a disease of sheep in Great Britain and other countries of Western Europe more than 250 years ago, scrapie has been reported throughout the world. Only two countries are recognized by the United States as being free of scrapie: Australia and New Zealand.

The first case of scrapie in the United States was diagnosed in 1947 in a Michigan flock. The flock owner had imported sheep of British origin through Canada for several years. From this first case through August 1999, scrapie has been diagnosed in more than 950 flocks in this country.

Clinical Signs

Signs of scrapie vary widely among individual animals and develop very slowly. Early signs include subtle changes in behavior or temperament; these changes may be followed by scratching and rubbing against fixed objects—apparently to relieve itching. Other signs are loss of coordination, weight loss despite retention of appetite, biting of feet and limbs, lip smacking, and gait abnormalities, including high-stepping of the forelegs, hopping like a rabbit, and swaying of the back end.

An infected animal may appear normal if left undisturbed at rest. However, when stimulated by a sudden noise, excessive movement, or the stress of handling, the animal may tremble or fall down in a convulsivelike state.

Several other problems can cause clinical signs similar to scrapie in sheep, including the diseases ovine progressive pneumonia, listeriosis, and rabies; the presence of external parasites (lice and mites); pregnancy toxemia; and toxins.

The scrapie agent is thought to be spread most commonly from ewe to offspring and to other lambs in contemporary lambing groups through contact with the placenta and placental fluids. Signs or effects of the disease usually do not appear until 2 to 5 years after the animal is infected. Sheep may live 1 to 6 months or longer after the onset of clinical signs, but death is inevitable.

On the farm, veterinarians diagnose scrapie based on the appearance of its signs combined with knowledge of the animal's history. There is no officially recognized test for scrapie in live animals, although research is progressing in this area. Scrapie can only be confirmed by microscopic examinations of brain tissue at necropsy or by procedures that detect the presence of the abnormal prion protein.

Epidemiology

Scrapie is classified as a transmissible spongiform encephalopathy (TSE). The agent responsible for scrapie and other TSE's is smaller than the smallest known virus and has not been completely characterized. There are three main theories on the nature of the scrapie agent: (1) the agent is a virus with unusual characteristics, (2) the agent is a prion—an exclusively host-coded protein that is modified to a protease-resistant form after infection, and (3) the agent is a virino—a small, noncoding regulatory nucleic acid coated with a host-derived protective protein. The scrapie agent is extremely resistant to heat and to normal sterilization processes. It does not evoke any detectable immune response or inflammatory reaction in host animals.

Genetic variations among different breeds of sheep may play a role in whether sheep will become infected and how quickly clinical signs may appear. Researchers in Edinburgh, Scotland, identified a

gene, called scrapie incubation period (SIP), that controls the incubation period of scrapie in Cheviot and Swaledale sheep. Animals with short incubation alleles usually develop signs between 2 and 5 years of age. Sheep with long incubation alleles often die from what appear to be natural causes before the incubation period is complete. Because the incubation period can be longer than 5 years, it is not known to what extent or under what conditions infected sheep with long alleles might be able to transmit scrapie to healthy sheep. It is likely that the prion protein gene and the SIP gene are the same.

Further research involving additional breeds has suggested that genetic influence may not only affect incubation length but may also confer some degree of disease resistance.

The strain of the scrapie agent also appears to affect the development of clinical signs and the length of the incubation period.

In the laboratory, the scrapie agent has been transmitted to hamsters, mice, rats, voles, gerbils, mink, cattle, and some species of monkeys. There is no scientific evidence to indicate that scrapie poses a risk to human health. There is no epidemiologic evidence that scrapie of sheep and goats is transmitted to humans, such as through contact on the farm, at slaughter plants, or butcher shops.

Scrapie research efforts are currently focused on developing a practical live-animal test to diagnose infected sheep before they show signs, investigating transmissibility of the agent, identifying the scrapie agent and its different strains, identifying genes that influence scrapie infection, and examining the role of artificial insemination and embryo transfer in the transmissibility of the scrapie agent. Recent research studies using experimentally infected sheep suggest that embryos may play a role in the spread of scrapie.

Related Diseases

The TSE family of diseases includes bovine spongiform encephalopathy (BSE), which affects cattle; transmissible mink encephalopathy; feline spongiform encephalopathy; chronic wasting disease of deer and elk; and kuru, both classical and variant Creutzfeldt–Jakob disease, Gerstmann–Straussler–Scheinker syndrome, and fatal familial insomnia, five rare diseases in humans. TSE's have also been reported in Europe in captive wild ruminants, cats, and monkeys. The occurrence of TSE's in captive wild animals is believed to have resulted from BSE-contaminated feed.

Control Program

In 1952, the Secretary of Agriculture declared a state of emergency in an attempt to eradicate scrapie in the United States. Since then, the U.S. Department of Agriculture's (USDA) programmatic efforts to control scrapie have changed several times. The purpose of all past USDA programs was to identify scrapie and eradicate it. That approach changed somewhat with implementation of the Scrapie Flock Certification Program on October 1, 1992.

The voluntary program is a cooperative effort among producers, allied industry representatives, accredited veterinarians, State animal health officials, and USDA's Animal and Plant Health Inspection Service (APHIS). The program provides participating producers with the opportunity to protect their sheep from scrapie and to enhance the marketability of their animals through certifying their origin in scrapie-free flocks. In addition, APHIS regulations restrict the interstate movement of sheep from scrapie-infected and source flocks.

Operating an effective program to deal with this insidious disease will require cooperation among producer organizations, allied industries, and governmental regulatory agencies.

Additional Information

For more information about scrapie, contact

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Current information on animal diseases and suspected outbreaks is also available on the Internet. Point your Web browser to http://www.aphis.usda.gov to reach the APHIS home page.